SPONTANEOUS INTRACRANIAL HYPOTENSION

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INTRODUCTION

Spontaneous intracranial hypotension (SIH) is characterised by postural headache associated with low cerebrospinal fluid (CSF) opening pressure. The exact aetiology is not known and usually there is no history of dural trauma. The condition is seen more commonly in women than in men and the course is usually self-limiting.⁽¹⁾ We present a patient with the history of postural headache and the radiological features typical of SIH.

CASE REPORT

A 47-year-old man developed a sudden onset severe frontal headache while he was bending over in his garden. The headache was aggravated by standing and bending downwards always improved his symptoms within minutes. The headache was associated with nausea. He had a past history of paroxysmal atrial fibrillation. He presented to hospital the next day. Clinical examination was normal. An initial diagnosis of subarachnoid haemorrhage was considered. A computed tomography (CT) scan of the head was normal. Lumbar puncture revealed an opening pressure of 6cm CSF in the lying position, protein was mildly raised and xanthochromia was negative. A diagnosis of SIH was made based on the low CSF pressure and typical postural headaches. He was treated with an epidural blood patch with relief of his symptoms within two to three hours.

He was readmitted with postural headaches within three weeks. The headaches were much worse this time and there were signs of meningism. Magnetic resonance imaging (MRI) of the brain showed evidence of subdural hygromas and high signal of both frontal and parietal convexity along the tentorial margin and MRI of the spine was normal. He was transferred to the neurology unit where he had three further epidural blood patches which resolved his headache. However, he became ataxic and developed a change in personality and examination revealed cerebellar syndrome. A repeat MRI of the brain showed mild herniation of cerebellar tonsil. His case was discussed with the neurosurgeons who advised not to proceed to surgery. His speech and balance gradually improved and he was allowed home after two weeks. However, shortly afterwards he was readmitted with headache and collapse. On this occasion a CT scan of the brain showed bilateral subdural haematomas with mass effect which required drainage.

DISCUSSION

In 1938, Schaltenbrand described a syndrome of spontaneous positional headache with neck stiffness, vomiting, tinnitus and vertigo in patients found to have a low CSF pressure.⁽²⁾ Schaltenbrand called this condition spontaneous aliquorrhea

and postulated that it can be caused by occult dural tear (site of the leak uncertain but commonly the cervical spine), over absorption of CSF, or decreased CSF production. The incidence is estimated at 5 per 100,000 per year.⁽³⁾

SIH should be suspected in any patient with postural headache. The headache associated with SIH is probably caused by dilatation of the cerebral veins and meningeal vasculature, or it may be a consequence of the displacement of pain-sensitive structures secondary to low CSF pressure, or a combination of both factors.⁽⁴⁾ Other symptoms may include visual field deficits, transient visual obscuration, facial numbness, sixth nerve palsy, vertigo, tinnitus, nausea, vomiting, and, rarely, stupor, cervical myelopathy and Parkinsonism.⁽⁵⁾ Progressive personality and behaviour changes with memory loss also may occur.⁽⁶⁾ Although uncommon, coma has been reported as a presentation of SIH.⁽⁷⁾

Definition of SIH as proposed by the International Headache Society⁽⁸⁾

Diagnostic criteria of headache attributed to spontaneous (or idiopathic) low CSF pressure

- A. *Diffuse and/or dull headache that worsens within 15 minutes after sitting or standing, with at least one of the following and fulfilling criterion D:*
 1. Neck stiffness
 2. Tinnitus
 3. Hyperacusia
 4. Photophobia
 5. Nausea
- B. *At least one of the following:*
 1. Evidence of low CSF pressure on MRI (eg, pachymeningeal enhancement)
 2. Evidence of CSF leakage on conventional myelography, CT myelography or cisternography
 3. CSF opening pressure of less than 60mm H₂O in sitting position
- C. *No history of dural puncture or other cause of CSF fistula*
- D. *Headache resolves within 72 hours after epidural blood patching*

IMAGING

MRI scans of the brain and spine are the investigations of choice. If MRI scans of the spine are normal, radioisotope cisternography or CT myelography are performed to detect CSF leakage.

Typical brain MRI findings suggestive of SIH include the following:

- subdural effusions (10% of cases) caused by rupture of bridging veins are due to the decrease in CSF volume and downward displacement of the brain⁽⁹⁾
- diffuse thickening of the meninges (also known as pachymeningitis) and meningeal contrast enhancement⁽¹⁰⁾
- obliteration of basilar cisterns, descent of midline structures and flattening of the pons against the clivus⁽¹¹⁾
- mild tonsillar descent, one of the cardinal signs of SIH⁽¹²⁾

TREATMENT

Many cases resolve spontaneously without treatment. A conservative approach, such as bed rest, hydration, caffeine and use of an abdominal binder, has been found to be helpful in some patients.

The mainstay of treatment is injection of autologous blood into the epidural space, a technique known as blood patch. This is effective in relieving symptoms in about one third of patients, presumably by forming a dural tamponade, with a seal of the tear being achieved by the formation of a clot. However, there is a conceptual difficulty in understanding its effectiveness here as epidural blood patches are performed in the lumbar area, below the level of the termination of the spinal cord, whereas the typical lesion of SIH is believed to be at the cervical level. Many patients require more than one blood patch. Surgical repair of CSF leak is an option for those patients in whom a structural abnormality or focal CSF leak is identified.⁽¹³⁾

CONCLUSION

SIH should be considered in any patient with postural hypotension. The spectrum of clinical and radiographic manifestations is varied, with diagnosis largely based on clinical suspicion, cranial magnetic resonance imaging and myelography. Numerous treatment options are available, but much remains to be learned about this disorder.

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